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International Journal of Surgery Case Reports

journal homepage: www.casereports.com

More on 'No Gut Syndrome': A case report

Varun Jain^{a,*}, Sergio Huerta^b^a Maulana Azad Medical College and Lok Nayak Hospital, University of Delhi, Delhi 110002, India^b University of Texas Southwestern Medical Center, VA North Texas Health Care System, 4500 S. Lancaster Road, Dallas, TX 75216, United States

ARTICLE INFO

Article history:

Received 21 August 2015

Received in revised form 9 December 2015

Accepted 9 December 2015

Available online 17 December 2015

Keywords:

Short bowel syndrome

Near total enterectomy

Total parenteral nutrition

Severe mesenteric ischemia

Extensive bowel necrosis

ABSTRACT

INTRODUCTION: No Gut Syndrome refers to a condition which results after a near total enterectomy (NTE) has been performed. These patients are unable to sustain life without life-long parenteral nutrition (PN). Re-establishment of bowel continuity, complications, quality of life (QoL), and overall outcomes are important aspects to be considered in patients who might need a NTE. We have previously reported two similar cases as well as a literature review in patients with No Gut Syndrome. In the present report, we present an additional case and an update of the literature.

PRESENTATION OF THE CASE: A 62-year old man with multiple co-morbidities presented with features of acute small bowel obstruction. Exploratory laparotomy revealed severe mesenteric ischemia and extensive small bowel necrosis. Patient eventually underwent a NTE and was discharged on parenteral nutrition.

DISCUSSION: Near total enterectomy (NTE) is a clinical condition in which a patient is left without or with minimal length of small bowel along with either gastrointestinal continuity or exteriorization of the proximal end. This condition has remained a dilemma for surgeons worldwide chiefly as a result of its perceived incompatibility with life. There are only a few available options for proceeding with treatment, however maintenance on long term parenteral nutrition has shown promising results.

CONCLUSION: Long term parenteral nutrition serves as a viable treatment option for managing patients after a NTE.

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1. Introduction

Short bowel syndrome (SBS) refers to the clinical entity resulting from loss of a significant portion of small bowel absorptive surface area due to surgical resection or bypass. The length of the resected bowel left in place that might lead to SBS is dependent on several factors including: portion of bowel resected (proximal vs distal), overall health of bowel, presence or absence of ileo-cecal valve, history of prior radiation therapy and ability to maintain adequate oral nutrition [1]. This syndrome is characterized by maldigestion, malabsorption, and malnutrition. The nutritional status of a patient is dependent on adaptation in the remaining bowel and a combination of pharmacologic and nutritional therapies.

Near total enterectomy (No Gut Syndrome), on the other hand, is considered a step beyond SBS and refers to the removal of almost the entire small bowel usually from the ligament of Treitz till the ileo-colic junction but may even extend up to the transverse colon. This condition arises uncommonly as an unfortunate consequence of extensive vascular compromise resulting in extensive bowel necrosis. As opposed to SBS, this entity leaves the patient without

sufficient length of bowel to undergo adaptive changes and, therefore, these patients must rely on life-long parenteral nutrition (PN). We have previously reviewed the literature on this condition [2]. However, we continue to observe similar cases, both, in the literature and in our clinical practice. This case reviews a patient with a similar situation after undergoing a near total enterectomy due to superior mesenteric ischemia and being successfully maintained on parenteral nutrition for two months.

2. Case report

This case report was undertaken after careful review of records following CARE criteria [3]. A 62 year old man had presented to the emergency in January of 2015 with signs and symptoms suggestive of small bowel obstruction. His past medical history was significant for an episode of ischemic colitis eventually resulting in a cecal perforation for which a right hemicolectomy with ileo-colonic anastomosis and a diversion loop ileostomy had been performed a year prior to presentation. A vascular surgery consult at that time, had determined him to be an unsuitable candidate for revascularization due to severe calcification of almost the entirety of his mesenteric vessels. He also had a history of COPD as well as multiple vascular interventions performed on bilateral lower extremities previously.

* Corresponding author.

E-mail address: varunjain7@hotmail.com (V. Jain).

At this admission, his SBO improved over the initial two days of conservative management, but then deteriorated with new onset diffuse abdominal pain and distention. A CT scan was performed, which revealed pneumatosis intestinalis (air within the bowel wall) throughout the small bowel. An exploratory laparotomy was performed and it revealed inter bowel adhesions with obstruction and a definite transition point at the region of dense adhesions, necessitating the resection of around 50 cm of small bowel and take down of the ileostomy leaving small segments of healthy bowel and approximately a 140 cm segment that was severely compromised. The bowel was left in discontinuity and planned for a second look after 24 h.

Second look revealed necrosis of almost entire length of small bowel with doubtful viability of 35 cm proximally and 15 cm distally. Resection of the necrotic bowel was performed (218 cm) and again the bowel was left in discontinuity, planned for another look the next day. At this point we were faced the dilemma of whether to proceed with further resection if the need arose or to simply refrain from any further radical treatment.

After detailed discussion with the family members regarding the patient's prognosis, operative mortality risk and expected post operative complications and considering our past successful experiences with total parenteral nutrition, the decision was made to proceed with any necessary treatment. Therefore, on re-exploration the next day, distal part of the proximal jejunum was found to be devitalized and was resected. A hand-sewn jejuno-ileal two layered anastomosis was performed with around 10 cm of the viable terminal ileum.

Intervention details and post-operative progress was assessed by reviewing medical records. In the immediate post-operative period the patient recovered well and PN was initiated on POD2. The only complaint was a high volume watery diarrhea which improved within 4 to 5 days after initiation of Octreotide. On POD5 the patient developed wound dehiscence with an entero-cutaneous fistula which was managed by a wound vacuum as well as wet-to-dry dressings. The patient tolerated PN well with BMI stabilizing around 18 kg/m² after 10 days, as compared to his pre-operative BMI of around 22 kg/m² and liver functions remaining normal even after almost 1 month on PN. He had marked improvement over the course of his hospitalization and on POD #30 he was discharged to a nursing facility near his house. A recent review of the records indicated that the patient had passed away at the outside facility. The cause of death was unknown.

A review of the literature identified an additional case, that describes a 78-year old woman who underwent a near-total enterectomy with end to end anastomosis as a result of extensive necrosis due to deep abdominal vein thrombosis [4]. Post-operatively this woman had developed features suggestive of bowel obstruction. A second look operation revealed a rather unusual cause. There was a large intraluminal clot at the site of the anastomosis which probably resulted from massive bleeding due to the continuous heparin administration after the first operation. After the second operation the patient improved clinically, was discharged on parenteral nutrition, and was noted to be doing well at 5 months follow up.

3. Discussion

Near total enterectomy continues to be an extremely important surgical issue, which is likely underreported in the literature. There are many conditions causing near total small bowel necrosis with devascularization being the common terminal event [5,6]. NTE is typically considered to be a lethal condition as many patients suffer from the cardiac insults that lead to the bowel ischemia. However, where there is hemodynamic stability and the patient along with

the patient's family desire to continue with treatment, one of the dilemmas regarding the next steps in management include placing these patients on permanent PN. Associated complications of permanent PN include hepatobiliary, electrolyte disturbances and line sepsis related, and have been previously reviewed [7]. These however, are surprisingly uncommon allowing patients to live much longer on PN.

Without resection a 100% mortality would be expected from the septic complications associated with the necrotic bowel. There are two broad issues in the operative decision making for patients undergoing a NTE. These are chiefly dictated by the overall status of the patient and wishes of the patient as well as the family. Therefore, one option is palliative care, and the second option being salvage. With all the advancements in medical science and visceral transplant, the experience and success with small bowel transplantation has only been limited [8,9]. Currently there are defined indications for SB transplantation, few of which include intestinal failure due to liver disease, inability to maintain venous access, recurrent catheter related sepsis (especially fungal), frozen abdomen, complete porto-mesenteric thrombosis or involvement of hepatic hilum or root of mesentery by tumor [9]. While age is not one of the factors, it must be considered in the overall decision making algorithm (the patient in the present report was 62 years-old, but with a burden of co-morbid conditions as a result of atherosclerosis).

NTE with permanent PN has been reported to have excellent outcomes. We therefore feel, this entity needs to continue being reported so that previous results and subsequent decisions can be discussed with the patient and patient's family should this scenario occur in clinical practice. We have previously reported two cases and a review of literature on patients who have undergone a NTE and have been successfully maintained on PN [2]. Since then we have managed an additional case that has been reported here in a similar manner and found an additional citation in the literature [4].

Long term maintenance on PN is not without its morbidity. However, these are less than expected and most patients outlive these complications. Outcomes are comparable to a SB transplant [7]. QoL is also comparable [10,11] and in some cases even better than in those with intestinal transplantation [12,13]. Another issue that remains to be addressed is cost. PN is one of the most expensive outpatient treatment modalities that we provide today and the financial implications need to be addressed prior to initiation.

Complications, QoL, and costs are all factors that result in a clinical dilemma while proceeding with further intervention in patients requiring a NTE. We faced a similar dilemma in the patient presented in this case. In such a situation, the only viable option was radical surgery for which the outcome seemed like one with multiple morbidities yet it was the only one with a chance for survival. The reason for performing an anastomosis was to maintain gastrointestinal continuity so that the fluid/electrolyte disturbances and morbidity associated with a stoma could be avoided. These patients with an anastomosis of the duodenum or proximal jejunum with the colon do have a defined set of problems the most prominent of which is recurrent episodes of watery diarrhea [14,15]. However, it has been observed that with the passage of time these episodes gradually reduce in frequency as well as severity [15,16]. Our previous experiences along with our experience in this case as well, have been encouraging in this regard [2]. Other options are also available, but have not been as successful. Those available are exteriorisation of the duodenum and colon if sufficient length is available. This however, is associated with massive fistula outputs leading to marked fluid and electrolyte imbalance necessitating an anastomosis at a later date [14,17]. Alternatively creation of a jejuno-gastrostomy [17] or a duodeno-gastrostomy

[18] along with placement of permanent gastric tubes have also been reported.

With only limited documentation of such cases in the literature, there is obviously a lack of conclusive evidence or comparison between the different modalities available to manage such massive bowel necrosis. However, from the review of literature compiled in our previous report and from our own experience, we have sufficient evidence to support NTE in patients with hemodynamic stability who wish to proceed with further treatment. NTE remains a realistic and preferable initial choice for cases with massive small bowel necrosis and these patients can then successfully be maintained on life-long PN.

4. Conclusion

Recent experience with this case as well as from the two cases described by us earlier re-asserts that in this seemingly rare and otherwise fatal clinical condition if the patients overall hemodynamic status is favorable and the patient/family wish to proceed with treatment, near-total enterectomy with end to end anastomosis and long term maintenance on parenteral nutrition remains a viable solution and provides such patients with a respectable chance at survival on PN.

Conflict of interest

All authors declare that there is no conflict of interest.

Funding

There are no study sponsors.

Ethical approval

Ethical clearance obtained.

Patient consent

Written informed consent was obtained from the patient for publication of this case report.

Author contribution

Varun Jain—Data collection, writing the paper, literature search.
Sergio Huerta—Patient management, study concept, writing the paper.

Guarantor

Sergio Huerta, MD, FACS.

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